The history of portal hypertension

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When studying the history of portal hypertension (PH)[1] - one is impressed, as in all branches of medical history, by the inquisitive spirit and the intuition of early investigators who, with simple technical means, acquired a surprising degree of knowledge and understanding. Thus, as early as 1543, Vesalius[2] drew an anatomical picture of the portal venous system (Figure 1) to which not much has been added. In the 1650s, only 25 years after Harvey's discovery of the blood circulation, Glisson[3] at a dissection in London, established the portal vein as the vessel by which blood was collected from the gastrointestinal tract and returned to the systemic circulation. His tool was a goose quill which he inserted into the portal vein and through which, using a pig's bladder, he injected milk liverward, observing how the liver turned pale as the milk traversed it on its way to the vena cava and finally to the lungs.

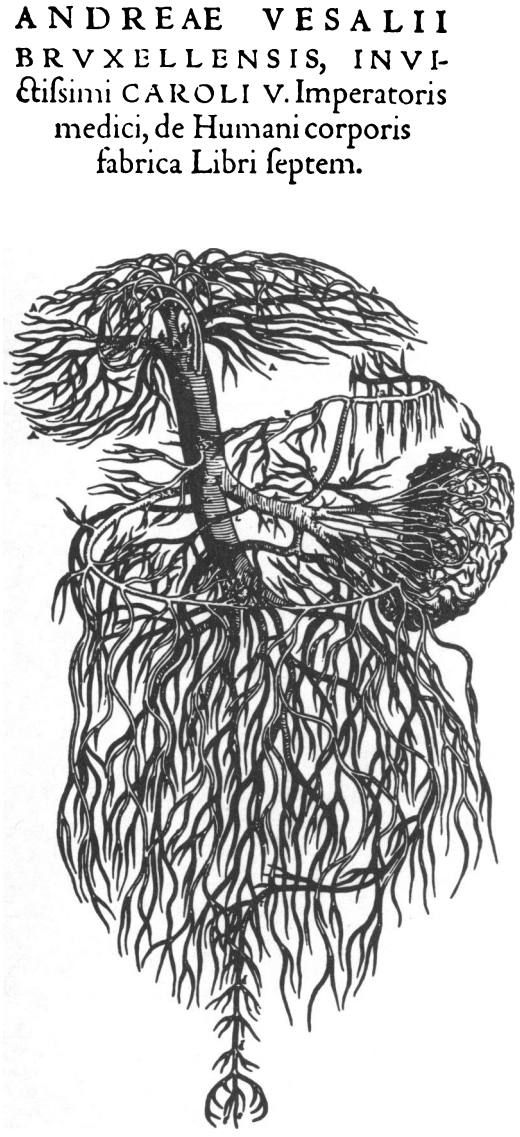
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Figure 1. Vesalius's pictorial of the portal venous system

Vesalius had already touched on the core of the pathophysiology by describing a case of bleeding haemorrhoids and suggested that this was due to a dilatation of the portal branches. The suspicion that gastrointestinal haemorrhage could be caused by the derangement of the portal circulation was passed on, two centuries later, by Morgagni[4]. He described a patient, observed by his teacher Valsalva, who had died from gastrointestinal bleeding and was found to have 'polypoid concretions' in the splenic vein and dilatation of the short gastric veins.

During the nineteenth century it became increasingly clear that the clinical picture of splenomegaly, ascites, and gastrointestinal haemorrhage generally was due to obstruction to the flow in the portal system. Cases were described by Puckelt[5] in 1818 and by Cruveilhier[6] in 1832.

In 1841 Raciborski[7] specified that collaterals could develop between the portal and the caval system via the short gastric veins, haemorrhoidal veins and veins in the abdominal wall. Later Sappey[8] added oesophageal varices as another link. Oré[9] commented that the portal vein often was occluded by a thrombus in patients dying from cirrhosis. Dusaussey[10], who in 1872 wrote an important thesis 'Studies on oesophageal varices in liver cirrhosis', believed that the obstruction to portal flow was a consequence of liver cirrhosis.

Banti's conception that the syndrome carrying his name was caused primarily by disease of the spleen[11] was shown to be erroneous by Warthin[12] in 1910 and now the term, Banti's disease, is only of historical interest.

The term portal hypertension, now universally accepted, is a child of this century, introduced by Gilbert[13] in 1902. However, it was not until 1937 that Thompson[14] could verify the increase in portal pressure directly during laparotomy. In 1950 Davidson[15], without opening the abdomen, obtained a 'pressure close to that in the portal vein' by puncturing one of the dilated abdominal wall veins (the Caput Medusae). Another non-invasive measurement was described in the following year by Myers[16] and by Friedman[17], who found that in liver cirrhosis the portal pressure could be estimated by occlusive catheterization of a hepatic venule. Finally, in 1953, Lebon[18] of Algeria with the knowledge from McNee[19] that there is open circulation between the splenic pulp and the splenic vein, diagnosed PH by percutaneous measurement of the intrasplenic pressure. At the same time they obtained excellent spleno-portographic images after injection of contrast material into the spleen.

In 1900, Preble[20] had collected 60 cases from the literature of liver cirrhosis where the patient had died

from haematemesis. He found that 80% of them had oesophageal varices. It was on these that therapeutic efforts were first concentrated since haemorrhage from these varices is the most dangerous, and often fatal symptom of PH. Westphal[20] was far ahead of his time when, in 1930, he published his method of temporarily stemming the bleeding by compression of the varices with a rubber balloon attached to a double lumen gastric tube. However, it was not until 1947 that it was introduced in the USA by Rowntree[22].

More lasting results were obtained when Crafoord[23] injected sclerosing solution around the varices via oesophagoscope. Crafoord, a famous pioneer in cardiac surgery, was also the first to use heparin for thromboembolism. Like Glisson, Crafoord is an example of how a great spirit can often make several imprints on developments. The method fell into oblivion for decades but was later re-established as one of the main therapeutic tools in PH.

It was always felt that local measures were unsatisfactory as the PH persisted, exerting its injurious effects on the splanchnic organs, causing splenomegaly, ascites, congestive gastritis, etc. The increasing understanding of the chain of events (portal obstruction; PH; splenomegaly; dilating collaterals; fatal haemorrhage) stimulated a search for a more radical approach aiming at reduction of the portal pressure.

Two roads seemed open: either reducing the blood-flow into the system or increasing the outflow. Inflow reduction through splenectomy gave only temporary relief and the attention was therefore directed towards ways of enhancing outflow. The healing efforts of Nature itself have been strikingly ill-fated in this field. When substituting degenerating liver parenchyma with the usual fibrous tissue, the portal flow gets strangulated, and when she looks for a way out, helped by the pressure gradient expanding the collaterals, the dangerous oesophageal varices are produced. The risk of haemorrhage from these vulnerable varices makes it essential for surgery to find other pathways.

The Dutch surgeon, Talma[24], had observed the rich network of small collaterals in peritoneal adhesions, connecting the portal and the caval systems, and suggested increasing them by sewing the omentum to the abdominal wall. This was first tried by his pupil Lens[25] in 1882. The new collaterals turned out to be entirely inadequate and the method was soon abandoned. In Britain the operation was known by the name of Talma-Morison.

During the last half of the nineteenth century it was commonly believed that animals died promptly if their portal vein was suddenly occluded.

The sturdy medical individualist Nicolai Eck of Russia was highly skeptical of the convictions of the day and set about proving that the dog, at least, could survive sudden and complete portal occlusion provided portal blood flow was simultaneously diverted into the vena cava[26].

In his article, in 1877, Eck[27] stated:

I am conducting these experiments with the purpose of clarifying some physiologic problems as well as to determine whether it would be possible to treat some cases of mechanical ascites by means of forming such a fistula. I operated on 8 dogs, one recovered completely and lived in the laboratory for 2,5 months. Because of lack of attention, he ran away. I had to postpone further experiments because I was called to join the active army.

The Eck fistula was instead thoroughly investigated by a team in Pawlow's laboratory[28]. In 1893 they wrote 'As a keen surgeon, Eck claimed that he could release the obstructed flow as the portal blood could be diverted into the systemic circulation without danger to the organism'. The Pawlow school showed that this was not entirely true - their most important observation concerned a derangement of the protein metabolism, resulting in 'meat intoxication', corresponding to the encephalopathy in liver cirrhosis.

The portacaval shunt was used for the first time on a human being by Vidal[29] in 1903 (the anatomical conditions were unsuitable for the Talma operation that he had planned). The patient lived for 4 months without haematemesis. Vidal considered the bleeding and not ascites to be the main indication for the shunt.

Occasional shunts were reported with varying results, but it was not until 40 years later that the operation was crowned with uniform success. Thanks to important contributions from the Spleen Clinic at the Presbyterian Hospital (NY, USA), under the leadership of Allen Whipple[30]. Rousselot[31] studied the result of splenectomy in Banti's syndrome with PH. As the team members were discouraged by the results of the operation in liver cirrhosis, they concentrated their efforts on the more rational portacaval shunt. The work was promoted by two advances of which the most important was due to another Whipple, George H[32]. In a well-studied experimental series he proved that the hepatic artery alone is adequate to support relatively normal clinical activity for many years.

Another helpful contribution was Blakemores's[33,34] invention of a non-suture vascular anastomosis, intima-to-intima, over a vitallium tube. Allen Whipple reported proudly:

Using this method, Dr Blakemore and I have carried out ten of these major procedures, five consisting of uniting the splenic vein and the left renal vein, after removing the spleen and left kidney. In our last five patients we have anastomosed the portal vein to the inferior vena cava, end-to-side. All these patients have survived their operations. The result in five of them have shown such a marked improvement in their liver function tests and disappearance of ascites or hemorrhage that we have been encouraged to continue.

The high frequency of thrombosis in the anastomosis was the reason for the vitallium tube becoming obsolete when Blalock developed his ingenious everting matress suture. Whipple himself quoted a letter from Blalock, reporting four cases performed by him.

Whipple's report gave rise to a tremendous wave of enthusiasm for the shunt[1]. In 1954, however, McDermott[35] described a case of 'episodic stupor associated with Eck fistula in humans with particular reference to the metabolism of ammonia', and it became evident that Pawlow's 'meat intoxication' occurred as post-shunt encephalopathy in a considerable number of patients.

This complication has continued to cloud the outlook, despite Warren's[36] endeavours to save part of the portal flow through the liver with his peripheral spleno-renal shunt. The frequency of the complication has stimulated more conservative direct attacks on the bleeding varices, and sclerosing therapy has become a prominent alternative to the shunt. Nearly 50 years later, there is still good reason to quote

Whipple: 'The problem of therapy for hemorrhage in cirrhosis will continue to be a serious one'.

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